

GUILLAIN-BARRÉ SYNDROME

RATIONALE FOR PHYSIOTHERAPY MANAGEMENT OF THE ACUTE SEVERE PATIENT

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Introduction

The Guillain-Barré syndrome (GBS) is characterised as an acute, symmetrically progressive, inflammatory polyneuropathy¹. The Guillain-Barré syndrome is one of the most common forms of polyneuropathy. The occurrence rate is 0.6 to 1.9 cases per 100,000 population per year. According to Beale and Miller it is estimated that approximately one new case of GBS is seen per week at Baragwanath hospital².

The syndrome was first described in 1916 by Guillain, Barré and Strohl¹. In the years that followed reports of numerous series of patients were published, often with pathological studies, that gradually built up the clinical picture. After the swine flu incident in 1976 and 1977 in North America, a committee was set up to determine the criteria for diagnosis of Guillain-Barré syndrome. Their findings were published in 1978³.

The classic presenting features are progressive motor weakness of more than one limb, areflexia (loss of tendon jerks) and mild sensory symptoms or signs³. Electrodiagnostic studies are particularly valuable in the differential diagnosis, and typically show an evolving pattern of multifocal demyelination⁴. After the first week of symptoms a raised spinal fluid protein level without pleocytosis is also strongly supportive of a positive diagnosis³.

Clinical criteria, spinal fluid protein elevation without cells and nerve conduction abnormalities are the mainstay of diagnosis.

The criteria for diagnosis of GBS and Chronic Inflammatory Demyelinating Polyneuropathy (CIDP) are very similar. They are distinguished by a different time course. CIDP is progressive over a period of more than two months⁵. The course of CIDP may be either relapsing and remitting or a subacute onset followed by steady progression or subsequent remission⁶. The aim of this paper is to review the aetiology, pathology, clinical features and treatment of GBS with particular reference to the physiotherapeutic management of the acute severe GBS patient.

Aetiology

The syndrome may occur in either sex and at any age, with a minor peak frequency in young adults and a second, larger one in the fifth to eighth decades of life⁴. Over half of Guillain-

Barré syndrome patients experience symptoms of viral respiratory or gastrointestinal infections in the 1-3 weeks prior to the onset of neurological symptoms⁷. An auto immune basis for the Guillain-Barré syndrome seems probable, however, with the diversity of triggering agents it seems unlikely that a single antigen is involved in the origin of GBS⁸.

Pathology

The main pathologic feature of GBS is segmental demyelination of the peripheral nerves. Some Wallerian degeneration may occur⁸. Axonal involvement may be so severe that it has been questioned whether a primary axonal form of GBS exists⁵. Fibres with more myelin (motor and those sensory nerves carrying proprioceptive information) are usually more severely affected, correlating with the signs of weakness and diminished vibration and joint position sense in GBS. The more thinly myelinated axons, which supply sensations of cutaneous pain, touch and temperature are less involved, apparently explaining early sparing of these systems in GBS⁹.

Clinical Features

Classic GBS has an acute onset of minor sensory changes such as a dysaesthesia in the feet, less often in the hands, followed by rapidly ascending weakness. Half the patients experience pain which may be present from the onset and be severe. The pathophysiology of the pain associated with GBS is poorly understood. In 1984 Ropper *et al*, analysed the clinical features of pain in patients with GBS. Their results suggest that alterations in muscle related to neurogenic changes may cause the typical pain of GBS¹⁰. Ropper later suggested that the pain and paraesthesias are probably caused by spontaneous discharges in demyelinated sensory nerves¹¹. Connelly *et al* (1990) considered the pain to be of two distinct types. One is a deep pain generally maximal in the back and buttocks and lower extremities ("the typical pain"). This is associated with tenderness and pain with passive movement of the affected muscle groups. Distinguished from these are symptoms that resemble causalgia with hyperaesthesia and a constant burning sensation in the extremities¹². Superficial sensory deficit, when present, is always of the "glove and stocking" type.

Muscle weakness usually starts in the legs and ascends to the arms. Proximal muscle weakness may be prominent from the outset^{7,11}. The weakness is fairly symmetrical and usually involves the trunk muscles.

The clinical course of the disease can be divided into three parts. The progressive phase lasts from the day the first motor or sensory symptom becomes apparent until progression stops. This usually occurs within three weeks. During the plateau phase which follows, the clinical signs remain the same (mean duration 10-14 days¹³). Duration of the recovery phase varies widely. Patients with severe motor weakness have a long recovery phase lasting from six to 24 months or more. Those patients who become bed bound and ventilator dependent within five days tend to have the most prolonged disability and may develop severe permanent weakness¹⁴.

Cranial nerves particularly facial (VII), oculomotor (III), and bulbar (IX to XII), may be affected causing facial weakness, diplopia and difficulty in swallowing respectively⁹. Respiratory failure of sufficient severity to require assisted ventilation occurs in one quarter of patients, although milder degrees of respiratory muscle involvement is much more common⁷.

ABSTRACT

The critical importance of supportive care including physiotherapy to the acute severe Guillain-Barré patient is emphasised in numerous reports in the literature. However, there are no systematic studies of physiotherapy in Guillain-Barré syndrome (GBS). This article examines the aetiology, pathology, clinical features, prognosis and treatment of GBS with reference to the physiotherapists' management of the acute severe patient treated in an intensive care unit.

Autonomic dysfunction occurs frequently in GBS. Manifestations include a wide range of cardiac arrhythmias, lability of blood pressure regulation, abnormal haemodynamic responses to drugs, ECG abnormalities, pupillary dysfunction, sweating abnormalities, urinary retention and gastro-intestinal dysfunction⁴. The presence of autonomic neuropathy cannot be predicted from the severity of the motor or sensory deficit¹⁴.

Like any patient in the ICU, the patient with severe GBS may exhibit a number of mood states, including demoralisation, sadness, fear and anxiety. Patients may also suffer from feelings of hopelessness and isolation⁴.

According to the literature 14% to 23% of all GBS patients go through a period of agitation and mental confusion. When other causes have been excluded this can be considered as an intensive care psychosis. It is caused by a combination of the patients anxiety, lack of sleep due to the intensive care environment, continuous pain and sensory deprivation as a result of disturbed sensory input. The signs usually subside in 1-2 weeks either spontaneously or with the aid of sedative drugs¹⁵.

Outcome

Most patients with GBS will make a good spontaneous recovery if they receive competent supportive treatment. The factors predictive of poor outcome with slow recovery or permanent disability are: age over 60 years, development of severe paralysis within five days of the onset, respiratory failure requiring ventilation, and mean distal compound action potentials of less than 20% of normal⁷. Another prognostic factor is the duration of the plateau phase. Patients with a plateau duration of more than 18 days have more residual signs¹⁶.

The mortality rate in severe GBS is 8-10%. Of the survivors 60% will make a full recovery, the remaining 40% will show some permanent symptoms and signs eg. distal leg weakness, distal sensory loss, absent ankle-jerks¹⁷. In a study by de Jager and Minderhoud in 1991, the most serious residual paresis was found in muscles innervated by the median, ulnar and peroneal nerves and distal branches of the tibial nerve. These nerves all pass along an entrapment site: the carpal tunnel, the ulnar sulcus, the fibula head and the tarsal tunnel. They suggested the entrapment may be contributory to the development of residual motor abnormalities¹⁶.

Treatment

Because of the severity of the disease effective treatment has eagerly been sought over the years. Major clinical trials have been conducted recently focusing on the value of corticosteroids, plasma exchange (PE), and high-dose immune globulins (IgIV) in the treatment of GBS and CIDP.

In 1995 van der Meche and van Doorn concluded that GBS patients with considerable deficit resulting in difficulties with locomotion should be treated with IgIV or alternatively with PE as soon as possible during the active phase of the disease (ie., in the first two weeks, or beyond that in the small percentage of patients showing ongoing progression of the disorder)⁵.

In the acute progressive phase the severe GBS patient is closely monitored in an ICU. The main concerns are respiratory failure and autonomic dysfunction. Electrocardiographic monitoring allows prompt recognition and treatment of arrhythmias. Deterioration of vital capacity to less than 15-20 ml/Kg (a vital capacity of approximately 1 litre in a 65 kg adult) and inability to clear secretions because of bulbar paresis and weak cough are indications for elective intubation. The optimal position for measurement of vital capacity is with the

head of the bed at about 60 degrees, as this allows maximum lung expansion without pushing the diaphragm upwards⁹. In cases of facial weakness it is necessary to hold the lips gently around the mouthpiece to ensure a seal. The nose should be pinched closed as well.

Current practice suggests that tracheostomy should be performed if intubation is required for two weeks or longer⁹. Nasogastric tube feeding should be used for patients with bulbar dysfunction.

Subcutaneous heparin and elastic stockings are used to prevent deep vein thrombosis and pulmonary embolism⁷.

Virtually all deaths and morbidity in GBS are due to secondary complications⁹. Chest physiotherapy (postural drainage, vibrations and sterile tracheobronchial suctioning) and frequent positional changes are very important care factors in GBS. Inflamed nerves are more prone than usual to secondary pressure damage and particular care must be taken to avoid pressure on the fibular head (peroneal nerve), medial humerus (radial nerve), or medial elbow (ulnar nerve) as these are related to most of the residual neurologic deficits in GBS patients⁹.

Mobilisation of the nervous system has recently emerged as an adjunct to the treatment of pain syndromes. The nervous systems mechanical and physiological functions are interdependent²¹. There is some evidence (and it is the authors experience) that increased pain and abnormal neural tension signs occur on testing patients with GBS^{20,24}. It is suggested that an awareness of neural tension signs may assist physiotherapists in positioning of patients to avoid or relieve discomfort caused by tension on neural structures, particularly in the acute stage²⁰.

Taking pain and neural tension into consideration, positioning and passive movements are performed to prevent the effects of immobilisation on the muscles and joints. When muscles are immobilised in the shortened position they lose sarcomeres in series from their muscle fibres and changes occur in muscle connective tissue resulting in increased muscle stiffness²⁵. The gastrocnemius and soleus muscles and the long finger flexors are particularly susceptible to developing contractures. In addition, periarticular connective tissue changes and the development of intra-articular adhesions occur with immobilisation which will limit joint range. Care must also be taken to prevent overstretching of muscles and joints during positioning and passive movements.

For pain relief most patients will respond to analgesics or non-steroidal anti-inflammatory drugs eg. Ibuprofen. However, Genis *et al* found that although many drugs have been used as analgesics in GBS including Quinine and oral and parenteral opiates, it has been difficult to achieve rapid and lasting pain control. In their study they satisfactorily relieved pain in nine GBS patients using morphine epidural analgesics¹⁸.

Psychosocial issues should be addressed regularly. Effective communication is mandatory in patients who are physically helpless but mentally alert ie., on a respirator with a tracheostomy tube in situ. All patients must have some sort of call light or bell. Alphabet boards, lists of frequent requests and concerns posted above the bed and simple codes indicated by blinking or shrugging can aid communication.

Nursing care and technical examinations should be arranged and coordinated to allow sleep, rest and privacy^{7,19}. A room with radio, calendar, photos and other personal objects creates a familiar atmosphere and reduces psychological distress. Early education of both patient and family about the nature of the disease and its usually favourable outcome is also important⁴.

Conclusion

During the past decade, treatment of the GBS has changed from simple supportive care and complication management to an active process (plasma exchange and intravenous immunoglobulin infusion) that shortens the duration of illness, especially for those patients who are severely affected²⁴. The critical importance of supportive care including physiotherapy to the GBS patient is emphasised in numerous reports^{4,9,11,22,23}. However, there are no systematic studies of physiotherapy in GBS¹¹. This article has examined the current issues of GBS relevant to the physiotherapist's management of the acute patient. We need to investigate whether GBS patients develop typical postures and movement patterns as a result of immobilisation, pain or nerve damage. We do need to prevent trauma caused by adverse neural tension in the acute stage during positioning and passive movements but at what stage do we include neural mobilisation to prevent the negative effects of nerve entrapment? How do muscle recruitment patterns change with GBS and how does this affect our muscle strengthening programmes? These are some of the questions physiotherapists must answer in systematic studies to provide optimal therapy for our GBS patients.

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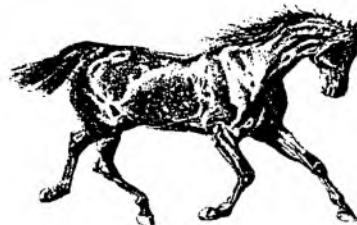
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Ninth International Therapeutic Riding Congress

Sponsored by the North American Riding for the Handicapped Association (NARHA), the Ninth International Therapeutic Riding Congress will be held July 14 - 20, 1997, in Denver, Colorado.

Held every three years, the congress provides an opportunity for professionals from around the world to share advances made in therapeutic horseback riding for individuals with disabilities. A collection of papers and studies are to be presented at the congress. NARHA has received more than 70 papers from several countries, including Australia, Brazil, Canada, Germany, Great Britain, Israel, Italy, the Netherlands, New Zealand, Russia and the United States. A wide range of international attendees are expected, including riding instructors, volunteers, programme administrators, riders, therapists, medical professionals and other who share an interest in therapeutic riding.

Equine-facilitated therapy was developed in the early 1950's in Europe as a tool for improving the lives of individuals



with disabilities. Because horseback riding gently and rhythmically moves the rider's body in a manner similar to human gait, riders often show improvement in flexibility, balance, muscle strength and self-esteem. Individuals with virtually any type of physical, cognitive or emotional disability can benefit from therapeutic riding. Currently, more than 25 countries offer therapeutic riding activities in some form. In recent years, therapeutic riding activities have developed in Japan, Brazil, Israel and Croatia.

For more information contact:

- NARHA, P O Box 33150, Denver, CO 80233, USA. 1-800-369-7433, 303-452-1212. Fax 303-252-4610.

NARHA is a non-profit equestrian organisation founded in 1969 to promote and support therapeutic riding for individuals with disabilities. Currently, NARHA has approximately 500 member riding centres across the US and Canada, serving more than 28,000 individuals with disabilities.